

Catching congenital heart diseases early: the ten - point approach for Bhutan

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ABSTRACT

Congenital heart disease is the most common birth defect and is the main reason for out country referral of pediatric patients from Bhutan. Given the limited resources and expertise, detecting congenital heart defects is often delayed, resulting in delayed initiation of treatment and management of associated complications. Congenital heart diseases, if detected early, can be appropriately treated and complications minimized. A ten-point approach consisting of antenatal diagnosis, neonatal pulse oximetry, well child visits, sick child visits, immunization clinics, school health checkups, pediatric echocardiographers, pediatric cardiologists, basic echocardiography skills in pediatric residents and comprehensive birth defect surveillance is suggested, in order to catch congenital heart defects early in life.

Keywords: Birth defect; Congenital heart disease.

INTRODUCTION

Congenital Heart Diseases (CHD) is defined by Mitchell et al. as “a gross structural abnormality of the heart or intra-thoracic great vessels that is actually or potentially of functional importance”¹. They are the most common birth defects and the most common cause of infants deaths from birth defects worldwide². The overall incidence of congenital heart diseases range from 8-10 per thousand live births to up to 50 per 1000 live births. A study published in this journal by the author found an even higher incidence of congenital heart defects (68/1000 livebirths) in a referral hospital in eastern Bhutan³. CHDs are also the most common reason of referral of pediatric patients for tertiary care to hospitals in India. Without an established pediatric cardiac program and with a limited number of cardiologists in the country, providing optimum pediatric cardiac services is still a big challenge for the pediatric fraternity in the country. Even more challenging is the fact that congenital heart defects are not picked up early in life, denying many infants a proper diagnosis, optimum management and follow up plans.

Current Scenario

Resources are limited in our country for the early detection, management and follow up of children born with congenital heart diseases. The overall awareness of birth defects and congenital heart diseases is low in primary health care settings where the majority of our children are taken care of. The expertise to diagnose and manage CHDs is limited even at the level of district and referral hospitals. The recent initiation of recording and reporting of birth defects has created some awareness to a restricted number of staff and caregivers at the level of regional

referral hospitals. The birth defect surveillance may be able to give some data on the prevalence and proportion of CHDs from the overall birth defects in the country.

Currently, only three referral hospitals in the country have qualified pediatricians, and much of the diagnosis, care and follow up of cardiac patients is based in these referral hospitals. Echocardiography services are also limited to these three hospitals, and therefore the majority of children in the peripheries do not have an easy access to the standard diagnostic tools for detecting and following up congenital cardiac defects.

What can be done to catch congenital heart diseases early?

Being the most common of the birth defects, the most common reason for pediatric out country referrals and also one of the major causes of infant mortality from birth defects, CHDs deserve to be taken seriously. Detecting CHDs early can make a huge difference in their appropriate management and long term outcomes. The following outline is suggested to identify opportunities to increase the rate of detection of CHDs as early as possible so that a proper management and follow up plan can be instituted. The author would like to call this the “ten point” approach to detecting congenital heart defects early.

1. Antenatal diagnosis

The majority of women in Bhutan seek antenatal care. There is no data on the percentage of women who have antenatal fetal anomaly scans. This facility is available only at the National Referral Hospital which has a lone specialist in fetomaternal medicine. For most of the clients, antenatal scans are done to date the pregnancy, monitor the growth of fetus and to detect any major complications. The services are mostly offered by ultrasound technicians who do not have the requisite skills to do a thorough anomaly scan.

Congenital heart disease can be detected by a fetal echocardiography at an early gestational age of 17-19 weeks⁴. If the defect is suspected or diagnosed, the mother can be followed up or at least have the baby delivered in a referral hospital where

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echocardiography facilities can facilitate accurate diagnosis, commencement of treatment and referral if required. As referring all pregnant women for an anomaly scan to the referral hospitals is not practical, technicians at the district and regional level might need to be trained to pick up some of the most common and critical heart defects.

Further, pregnant women with risk factors like history of rubella infection, maternal diabetes, family history of CHD, alcohol use and use of certain medications like lithium, anti-convulsants and vitamin A derivatives, may be given the option of undergoing an anomaly scan for fetal heart defects even if it requires referral to the National Referral Hospital.

2. Neonatal Examination and Pulse Oximetry

This is the best and earliest opportunity to detect any congenital heart defects. All newborns delivered at various facilities should undergo a thorough clinical examination before discharge from the hospital. This should preferably be done by a pediatrician or a medical officer, giving special attention to the color, respiratory rate, femoral and peripheral pulses, careful cardiac auscultation and a pulse oximetry.

Pulse oximetry is now widely used in many countries as a screening tool to detect congenital heart diseases. It should be done before the neonate is discharged from the hospital and preferably after 24 hours of age. Oxygen saturation under 95% at 24 hours after birth has the best specificity⁵. Both upper and lower limb saturations should be noted. Any neonate with a saturation of less than 95% should be further examined for possible CHDs. Although this will detect only the cyanotic or critical lesions, it offers a good chance to pick up the more serious defects that will require a more detailed work up and early referral to tertiary care hospitals.

At the moment, Central Regional Referral Hospital in Gelephu offers pulse oximetry to all neonates before discharge. Although we have not detected CHDs through this, it serves to be reassuring to exclude the presence of any serious underlying cyanotic cardiac defects in the newborn. As part of the Comprehensive Birth Defect Action Plan, pulse oximetry (prior to newborn discharge) should be made mandatory for all institutional deliveries.

3. Well child visits

Thorough clinical examination with careful cardiac auscultation should be performed at all well child visits. If there is reasonable suspicion, simple chest x-ray can detect cardiomegaly, pulmonary plethora or oligoemia and guide us to a diagnosis of CHD. Many congenital heart defects especially those presenting early in the neonatal period or infancy, have a specific x-ray appearance and a fair guess can be made on the underlying defect even if Echocardiography facilities are not available.

4. Immunization clinics

Children attending Mother and Child Health (MCH) clinics should be assessed for color, fast breathing, head sweating or any

growth faltering and should be referred to a medical officer for appropriate evaluation.

5. Sick child visits

As usual, complete physical examination including the cardiovascular system should be done. Children with cardiac murmurs, growth faltering, recurrent pneumonias, recurrent wheezing, suspicious chest x-rays, feeding difficulties, head sweating, features of heart failure or family history of CHDs should be screened for CHDs preferably by an Echocardiogram.

6. School health visits

It is a regular practice to screen all school children for any health problems during school visits by health workers from the nearest hospitals and Basic Health Units (BHUs). Although schooling children are unlikely to have a serious underlying cardiac defect remaining undiagnosed, there are still a large number of children who are detected to have cardiac murmurs during school health check ups. It is quite possible for some of these children to have some of the less serious cardiac lesions like small Atrial Septal Defects (ASDs), small Ventricular Septal Defects (VSDs), bicuspid aortic valves, small Patent Ductus Arteriosus (PDA) or pulmonary stenosis. Even if asymptomatic, many of these lesions may either progress or may become symptomatic in adult life. Therefore, all school children with detected heart murmurs should be given the opportunity to undergo an elective echocardiogram to confirm or exclude a congenital heart lesion. Even if Echocardiogram is not possible, the child may be seen by a Pediatrician to ascertain whether the murmur is of clinical significance or not.

7. Training Pediatric Echocardiographers

Most of the echocardiograms performed in the country are by adult echocardiographers or adult cardiologists. Pediatric Echocardiography needs special skills and approaches that all adult echocardiographers might not be familiar with. It's thereby natural that many lessons may be missed or misdiagnosed. There is evidence of high incidence of diagnostic errors in pediatric echocardiograms performed in adult laboratories⁶.

It is therefore reasonable to propose for Pediatric Echocardiographers at least for the National Referral Hospital or train the existing adult echocardiographers on the special skills required in pediatric echocardiography.

8. Training Pediatric Cardiologists/Fellows

With increasing public awareness, improved health coverage, availability of qualified doctors even at the BHU level and the recent initiation of birth defect surveillance, it is likely that an increasing number of CHDs will be detected every year. The present practice of referring patients to India for cardiac surgery will continue until we have an established cardiac surgical program in the country. However, appropriate diagnosis,

management and follow up of pediatric cardiac patients including operated patients is best provided by Pediatric Cardiologists. It is high time for the country to have a few such specialists. This will also provide a back up for the adult echocardiographers who perform pediatric echocardiograms, in case of complicated or complex congenital heart diseases.

9. Echocardiography skills in Pediatric Residency Training

With the initiation of the four-year Pediatric Residency Program at JDWNRH, the nationwide shortage of Pediatricians will soon be a thing of the past. Larger district hospitals will probably start getting Pediatricians, promising a better health for our future generations. Incorporation of basic Pediatric Echocardiography skills in the upcoming Pediatricians will equip them with the capacity to perform basic echocardiograms, make appropriate diagnosis and institute accurate management plans. Simple echocardiography training protocols can be used and inexpensive portable echocardiography machines can be made available at larger district hospitals.

10. Comprehensive Birth Defect Surveillance Program

With the institution of birth defect surveillance and reporting in 2015, there is now a national birth defect registry which includes congenital heart diseases as one of the reported birth defects. The presence of one birth defect should alert the physician to look for other possible defects. For example, finding a cleft lip or palate should prompt the clinician to look for congenital heart defects as well. As CHDs are associated with many other common birth defects and syndromes, the pick up rate will definitely be higher if children with other birth defects are screened for congenital heart diseases.

CONCLUSIONS

Congenital heart diseases need to be detected early in order to initiate appropriate management, institute a proper follow up plan and avoid long term complications.

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